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Practitioner Review: Self-injurious behaviour in children with developmental delay

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Self-injurious behaviour in children with developmental delay

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Abstract

Background: Self-injurious behaviour is shown by a significant minority of children with developmental delay and has a substantial impact on child and carer wellbeing. Characteristics such as a greater degree of intellectual disability, autism spectrum disorder, some genetic syndromes and repetitive and impulsive behaviours are positively associated with self-injury. Prevalence generally increases with age into mid-adulthood and the behaviour is notably persistent.

Scope: In this review we discuss the dominant causal theory of self-injury which draws on the principles of operant learning. We evaluate the utility of this theory to account for all empirical observations of self-injury.

Findings: A model of self-injury is presented that extends a previous model described by Guess and Carr. The new model integrates child characteristics and operant learning principles in a phenotype x environment paradigm to explain the variance in developmental trajectory of the severity of self-injury.

Conclusions: Behaviour dysregulation, as evidenced by the associations between self-injury, self-restraint, repetitive and impulsive behaviours, is identified as potentially influencing the severity and persistence of self-injury. Risk markers for self-injury are identified and the extended model indicates points of intervention and highlights the possibility of risk related, targeted early intervention. The need for increased training of practitioners in the delivery of demonstrably effective interventions for self-injury is identified.

Introduction.

Self-injurious behaviour in children with intellectual disability and\or autism spectrum disorder (ASD) where intellectual disability is also present is an intractable and clinically challenging problem. Despite nearly 50 years of research there is little evidence that the most robust findings have been translated into widely available effective interventions or strategic initiatives. This inertia appears to be driven primarily by the lack of dissemination of clinical skills in assessment and intervention, the resource intensive nature of some interventions and the perceived limited efficacy of demonstrably effective interventions in the longer term. Here we provide an overview of the main themes that characterise the divergent research literature relevant to the assessment and treatment of self-injurious behaviour and describe a model of the development and persistence of self-injury that is consistent with the available evidence. This model provides a basis for identifying targets for clinical assessment and intervention at different stages of the development of self-injury at both a case and population level and highlights potentially productive research strategies.

Definition and conceptualisation

Murphy and Wilson's (1985) definition of self-injurious acts initiated by the individual that lead directly to physical harm remains useful today, with caveats regarding its use. The criterion of an observable outcome of the behaviour might contribute to underestimating the prevalence of self-injury in younger children which may have implications for early intervention and interpretation of prevalence data (see below). Also, using a criterion of outcome alone in prevalence or cohort studies does not recognise the potential importance of the form of self-injury. Head hitting, for example, is related to persistence of self-injury (Emerson et al., 2001) and a number of forms of self-injury occur at a higher prevalence in some syndromes than in contrast groups (e.g. lip and finger biting in Lesch-Nyhan syndrome; Christie et al., 1982; hand biting in fragile X syndrome; Symons et al., 2003; skin picking in Prader-Willi syndrome; Holland et al., 2003). These associations allude to different causal mechanisms and may be associated with different psychological characteristics.

Prevalence and persistence

Within the total population of people with intellectual disability estimates of the prevalence of self-injury vary from 4 to 24% (e.g. Cooper et al., 2009; Deb, Thomas, & Bright, 2001). Variability is related to the definition of the behaviour, the time window and sample characteristics. Studies investigating the prevalence of self-injury in children with intellectual disability are fewer in number and typically employ small samples and with limited robust data. A recent study in the UK of approaching 1,000 children with severe intellectual disability, generated prevalence figures of 17% for self-injury of any severity and between 4% and 5% for clinically significant self-injury (Oliver et al., 2012; Ruddick et al., In review). A systematic review of prevalence data shows that prevalence rises significantly with age up to approximately 30 to 40 years of age and decreases thereafter (Davies & Oliver, 2013). However, increase in prevalence below the age of 30 to 40 is not universal; Arron, Oliver, Moss, Berg and Burbidge (2011), for example, show this is not the case in Cri du Chat, fragile X, Prader-Willi, Cornelia de Lange, Lowe and Smith-Magenis syndromes. Similarly, Ruddick et al. (In review) report no significant difference in the prevalence of severe self-injury between children with severe intellectual disability under eleven years of age and those aged 11 to 18.

The limited available data suggest self-injury is very persistent. Taylor, Oliver and Murphy (2011) report approximately 84% persistence over 18 years, Emerson et al. (2001) 71% over 7 years and Cooper et al. (2009), using a definition of self-injury with a high threshold, 62% over 2 years. In combination, the majority of studies suggest that the prevalence of self-injury increases with age into adulthood and persists for many years. However, the association between age, persistence and individual characteristics (such as genetic syndrome or ASD), warrants further examination.

Child characteristics and behavioural correlates.

The first reference to behavioural phenotypes by Nyhan (1972) focussed on the possible association between Lesch-Nyhan and Cornelia de Lange syndromes and self-injury. Subsequently, numerous studies have sought to establish the prevalence of self-injury in syndromes and assess whether the prevalence is significantly higher than expected given degree of intellectual disability (the most well established correlate of self-injury).

Syndromes in which the prevalence of self-injury is higher than expected given relevant group characteristics include: Lesch-Nyhan, Cornelia de Lange, Cri du Chat, fragile X, Prader-Willi and Smith-Magenis, amongst others (Christie et al., 1982; Clarke & Boer, 1998; Collins & Cornish, 2002; Holland, Whittington, Webb, Boer & Clarke, 2003; Symons et al., 2003)

Arron et al. (2011) employed the same measure across syndromes and a contrast group and demonstrated a significantly higher prevalence of self-injury, but not necessarily aggression, in a number of syndromes. The dissociation between self-injury and aggression noted here and with age (see above) suggests different causes to the behaviours. Table 1 highlights some of the data on child and behavioural characteristics for which there is emerging evidence of an association with self-injury, including the genetic syndromes identified by Arron et al. (2011). Where available, odds ratios are presented to describe the relative odds of self-injury contingent upon the presence of these child and behavioural characteristics. The data from Arron et al. (2011) demonstrate that the presence of specific syndromes is associated with a 2 to 35 fold increase in the odds of self-injury. Importantly, these estimates are conservative as the contrast group of people with heterogeneous aetiology had a higher than usual prevalence rate of self-injury (26.8% vs. the typical estimate of between 4% and 24%).

One of the more robust findings in prevalence and cohort studies is that the prevalence of self-injury increases with degree of intellectual disability (Chadwick, Piroth, Walker, Bernard, & Taylor, 2000; Holden & Gitlesen, 2006; McClintock et al., 2003). As with age related prevalence there are exceptions (see Arron et al., 2011). Greater disability is associated with higher prevalence in Prader-Willi and Cornelia de Lange syndromes but not Fragile X or Cri du Chat syndromes. The prevalence of self-injury is not raised significantly in Angelman syndrome (a syndrome characterised by profound and severe intellectual disability) but is in Prader-Willi syndrome (in which moderate to mild intellectual disability is the norm) (Arron et al., 2011). Both exceptions warrant explanation.

A number of recent studies report an association between ASD with associated intellectual disability and self-injury with prevalence estimates ranging from 33 to 71%.

There is growing evidence that the prevalence of self-injury within ASD is higher than might be expected when degree of intellectual disability is controlled for (Richards et al., 2012). A meta-analysis of prevalence studies has shown that those with ASD are approximately six times more likely than those who do not have the diagnosis to show self-injury (McClintock et al., 2003). The association between degree of intellectual disability and prevalence seen in intellectual disability is evident in ASD (although the samples of those with intellectual disability are likely to include people with ASD). Within a number of genetic syndromes (Cornelia de Lange, fragile X and Down Syndromes) a higher score on a screening measure for ASD is associated with self-injury (Arron et al., 2011; Richards et al., 2012). This suggests that within groups at high risk for self-injury, ASD characteristics might add to or account for the risk.

There are numerous reports of the association between stereotyped behaviour and self-injury in prevalence and cohort studies and these have stimulated a number of interpretations (see Bodfish, Crawford, Powell, Parker, Golden & Lewis, 1995; Powell, Bodfish, Parker, Crawford, & Lewis, 1996; Rojahn, Matson, Naglieri, & Mayville, 2004). It is possible that this association can be accounted for by: 1) the association of ASD with self-injury, 2) the evolution of self-injury from stereotyped behaviours by selective operant social reinforcement (Guess & Carr, 1991; Oliver, 1993) or 3) a common underlying 'movement or movement control disorder' (Muehlmann & Lewis, 2012). In children with severe intellectual disability repetitive behaviour is associated with an increase in the presence of aggression and self-injury respectively of three and six fold with a four and sixteen fold increase in severe aggression and severe self-injury (Oliver et al., 2012). As the association between repetitive behaviour and self-injury is not unique, and severity in addition to presence only is predicted, these observations suggest that the association between these two behaviours cannot be accounted for simply by an evolution of a repetitive behaviour into a self-injurious one.

An increasing number of studies has identified an association between either impulsivity or ADHD and self-injury (Cooper et al., 2009; Bradley, Summers, Wood, & Bryson, 2004). As with ASD, Arron et al. (2011) showed higher levels of impulsivity in some genetic syndromes is associated with self-injury. Within ASD, Richards et al. (2012) and Richman et al. (2012) have shown the same association exists. These reports

are intriguing but warrant further examination using behavioural indices of impulsivity alongside caregiver report.

Behavioural correlates of self-injury that are frequently reported but rarely studied are self-restraint and the preference for imposed restraint (Powell et al, 1996). Early reports of these behaviours described children wrapping themselves in clothing, restricting the movement of hands and arms and showing a strong preference for wearing armsplints or headgear. These may not be uncommon (Oliver, Murphy, Hall, Arron & Leggett, 2003) and are of interest as they may have therapeutic value (see Powers, Roane, & Kelley, 2007) and suggest the behaviour might not be completely under control (see King, 1993). In Cornelia de Lange syndrome those who show self-injury and self-restraint have higher levels of compulsive behaviours than those who show self-injury but who do not self-restrain (Hyman et al., 2002). This association warrants investigation in other populations to evaluate if self-restraint and the preference for imposed restraint are associated with other behaviours normally considered indicative of compromised behavioural control, such as compulsive or repetitive behaviours.

A possible role for behaviour dysregulation

The association between repetitive behaviours, impulsivity and self-injury and the observation of self-restraint are of interest as they may help extend existing models of self-injury. The theoretical explanations of Turner (1997; 1999) of repetitive behaviour and, for example, of Nigg (2005) and Sonuga-Barke (2002) relevant to impulsivity, have identified deficits in executive functioning to account for observed behaviours. Turner has argued that as a result of specific cognitive impairments, the inability to modify or terminate ongoing behaviour accounts for the invariance and persistence of repetitive behaviour. Similarly, contemporary accounts of impulsivity in children with ADHD cite impaired inhibition of pre-potent responses and the inability to stop an ongoing response as a contributory mechanism. It is possible that the presence of repetitive behaviour and impulsivity are indicators of generally compromised behavioural self-regulation (via compromised executive function) and this would account for: 1) the association between repetitive behaviour and the severity, as opposed to just presence, of self-injury (see Oliver et al., 2012; as episodes of behaviour are initiated without inhibition and continue if there is no external intervention), 2) the

presence of self-restraint or the preference of imposed restraint (see Oliver et al., 2003; as forms of restriction of self-injurious acts are sought by the individual) and 3) the invariance of form and remarkable persistence of self-injury over years (see Emerson et al., 2001; and Taylor et al., 2011). Additionally, the explanation is compatible with the evidence from operant studies that self-injury can be evoked either when discriminative stimuli (environmental cues for the availability of reinforcement) and establishing operations (motivational states) are present (see below), or in response to pain because the explanation is focussed on the regulation of an established behaviour as opposed to the reason it might be initiated.

Low Mood and Self-Injury

In the literature on adults with intellectual disabilities it has been suggested that self-injury might be a 'depressive equivalent'. Evidence is, at best, tenuous and contested (McBrien, 2003; Tsiouris, Mann, Patti, & Sturmey, 2003; Davies and Oliver, 2014). The suggestion is of concern as there might be other important interpretations of an association between pervasive low mood and self-injury. Pain and discomfort (see below) is the most obvious reason that self-injury and low mood might be associated. Additionally, environments characterised by low levels of stimulation or coercive and punitive regimes might contribute to self-injury via operant mechanisms and simultaneously promote pervasive low mood and loss of interest in activities. Although the suggestion that self-injury is indicative of depression in children with intellectual disabilities has not yet gained ground, it should be considered extremely cautiously and, in clinical practice, only after pain and discomfort and environmental explanations have been considered, if at all.

Causes of self-injury

Operant learning

Operant learning theory accounts of self-injury propose that the behaviour is positively or negatively reinforced by sensory, tangible or social stimuli. More complete accounts identify the effect of self-injury (more specifically its short term cessation) on the behaviour of carers in a way that cultivates a mutual reinforcement cycle (Oliver, 1993; 1995). Evidence for this cycle comes from observational studies (Emerson, Hatton,

Robertson, Henderson, & Cooper, 1999; Hall & Oliver, 1992; Oliver, Hall, & Murphy, 2005).

Experimental evidence that self-injury can be a learned behaviour continues to expand with applied behaviour analytic studies demonstrating that: 1) self-injury can be evoked and rewarded by an increasing variety of environmental events, 2) self-injury can be reduced by manipulation of existing contingencies, 3) self-injury can be reduced by the introduction of adaptive behaviours that displace self-injury and 4) self-injury can be reduced by increasing the non-contingent availability of specific reinforcement. These experimental demonstrations support the argument that self-injury can be influenced significantly, favourably and unfavourably, by the immediate social and material environment.

Longitudinal naturalistic studies of self-injury in children have revealed that higher levels of self-injury when no social contact is available and greater concern about the self-injury on the part of carers, predict the future development of more frequent self-injury (Murphy, Hall, Oliver, & Kissi-Debra, 1999; Hall, Oliver, & Murphy, 2001a). Also, when the mutual operant reinforcement process is operative, self-injury is likely to increase over time (Oliver et al., 2005). These observations might be related, as concern might increase the likelihood of a socially reinforcing response by carers to effect short term cessation of self-injury that occurs when attention is not available and this is the nature of the mutual reinforcement process. Alternatively, concern might be heightened when compromised behavioural control by the child is evident as the behaviour is then more difficult to manage. However, these studies, and a similar study by Richman and Lindauer (2005), have demonstrated that in younger children with self-injury the social reinforcement process is applicable only to a minority of children. Further study of very young children who show self-injury is warranted.

The assessment of operant processes via functional analysis continues to be refined. Short analogue sessions within experimental functional analyses, variations of influential antecedents and reinforcers and the development of questionnaire methods have all helped to increase the validity of assessments whilst attending to ethical concerns. In this regard, studies that record precursor behaviours (behaviours that reliably precede episodes of self-injury) (e.g. Smith et al., 2003; Petty et al., 2009) are

of interest as they reduce the self-injury shown during assessment and identify the point at which adaptive responses might be reinforced. This is a promising area of research that could help to increase the effectiveness of applied behaviour analytic interventions.

It is clear from a number of studies that self-injury shown by children with genetic syndromes that are associated with self-injury might still be influenced by environmental events, even in Lesch-Nyhan syndrome (Hall, Oliver, & Murphy, 2001b). A productive line of research is the interaction between motivation related aspects of the behavioural phenotype of genetic disorders and operant reinforcement (Oliver, 1993; Langthorne, McGill, & O'Reilly, 2007; Tunnicliffe & Oliver, 2011; Langthorne, McGill and Oliver, 2014). Unusually strong motivation for social contact is evident in Smith-Magenis syndrome and has been shown to be related to self-injury by Taylor and Oliver, (2008), Sloneem et al. (2009) and Langthorne and McGill (2012). Similarly escape from social contact has been demonstrated as motivation for self-injury in fragile X syndrome (Hall, DeBernardis, & Reiss, 2006; Langthorne & McGill, 2012), Cornelia de Lange syndrome (Arron et al., 2006) and Rett syndrome (Oliver, Murphy, Crayton, & Corbett, 1993). This research is in its infancy but has the capacity to reconcile the apparently conflicting findings from the behavioural phenotype and operant literatures.

Pain and Discomfort

In the last decade a number of studies has emerged which indicate that pain might directly cause self-injury (see Symons, 2011). Luzanni, Macchini, Valade, Milani and Selicorni (2003) showed that gastro-oesophageal reflux was related to self-injury in Cornelia de Lange syndrome, presumably as a result of pain and discomfort. Breau et al., (2003) have shown that children with chronic pain self-injure near to the site of pain. Additionally, there is evidence that pain and discomfort can interact with environmental antecedents (Carr, Smith, Giacin, Whelan, & Pancari, 2003) to enhance motivation for operantly maintained self-injury. These studies extend the early observations that self-injury might begin as response to pain before being subjected to social reinforcement (see Carr & McDowell, 1980). In combination, these studies clearly indicate that assessment of pain as a cause of self-injury should be a clinical and research priority. More specifically, the association between pain, pain perception and

self-injury warrants investigation. Self-injury could moderate the perception of pain caused by ongoing health problems (Melzack & Wall 1965/1982; Woolf & Salter 2000). Additionally, compromised pain perception may influence self-injury. There is anecdotal and published evidence for a heightened pain threshold in some genetic disorders, such as Smith-Magenis, Prader-Willi and Cornelia de Lange syndromes, in which self-injury is prominent (Kline et al., 2007; Priano et al., 2009). Self-injury occurring for any reason might have lower response cost (in an operant conceptualisation) if the pain threshold is higher. However, a recent review of pain sensitivity in individuals with ASD reports that despite substantial anecdotal evidence of compromised pain perception, supportive experimental evidence is lacking (Allely, 2013). Instead it is posited that individuals with ASD may not express pain and discomfort in the same way as typically developing children, and thus a higher pain threshold is assumed due to the absence of pain related behaviours such as crying and comfort seeking. It remains critical therefore, to have knowledge of an individual's idiosyncratic pain behaviours or 'pain signature' in order to ensure that pain and painful health conditions are assessed and treated appropriately.

Movement disorder

An alternative cause of self-injury that has some empirical support but which receives less attention in the literature is the movement disorder hypothesis. This hypothesis is often a default explanation for self-injury that is invariant across environments and thus not immediately explicable within an operant framework. Evidence for self-injury as a disorder of movement hinges primarily on the association between self-injury and other movement disorders both within genetic syndromes and more widely, the effects of some psychoactive medication and animal models of induced stereotyped behaviours that result in injury (Gualtieri & Hawk, 1980; Lewis, Tanimura, Lee, & Bodfish, 2007; Stein, Niehaus, Seedat, & Emsley, 1998). In a recent review, Muehlmann and Lewis (2012) concluded that alterations in cortical basal ganglia circuitry underlie both self-injurious and stereotypic/compulsive behaviours. This shared pathophysiology could be an alternative explanation for the behaviour dysregulation described above. .

The use of medication as treatment for self-injury

Psychoactive medications are widely used, in up to 60% of individuals with intellectual disability, for the treatment of behaviour (Holden & Gitlesen, 2006; Tsiouris, Kim, Brown, Pettinger, & Cohen, 2012). A recent review highlights prescribing medication "off-label" (Farmer & Aman, 2013) to treat behaviour rather than psychopathology. Despite the widespread use of psychoactive medication, evidence for the efficacy of these medications to reduce self-injury is limited. Whilst some experimentally controlled trials have been conducted, many of these trials target global constructs such as 'irritability', rather than self-injury specifically. Therefore, whilst positive changes in irritability have been described for atypical antipsychotics including risperidone (e.g. Aman, De Smedt, Derivan, Lyons & Findling, 2002; Snyder *et al.,* 2002) and aripiprazole (Marcus *et al.,* 2009, Owen *et al.,* 2009), and combined treatments using risperidone and anti-convulsant medication topiramate (Rezaei *et al.,* 2010), it is not possible to use these studies as evidence for treatment of self-injury.

There are a limited number of controlled trials that have specifically measured changes in self-injurious behaviour through medication use. King *et al.,* (2009) published a controlled trial of the selective serotonin reuptake inhibitor (SSRI), citalopram, in individuals with ASD. The results showed no significant improvement in repetitive and restricted behaviours (including self-injury), but did show an improvement in irritability. The divergence in the outcomes for these categories of behaviour demonstrates the need to undertake studies that employ precise definitions of outcome variables. Other studies of SSRI efficacy produce equivocal results with some reductions in rate and frequency of self-injury in a limited number of participants (Lewis, Bodfish, Powell, Parker & Golden, 1996).

The most consistent evidence for medication use to treat self-injury comes from the results of naltrexone and naloxone trials. Controlled trials of the opioid antagonist naltrexone have typically demonstrated reductions in self-injury (Sandman, Barron & Coleman, 1990; Thompson, Hackenberg, Cerutti, Baker & Axtell, 1994; Symons *et al.*, 2001). This has led to a maintenance hypothesis of endorphin reduction in self-injury. This is supported by the observation that endorphin levels may be as raised following self-injury. It is possible that naltrexone and naloxone act by simply increasing the pain experienced from self-injury and hence influencing the response cost of an operant behaviour.

Overall, the evidence for medication use to treat self-injurious behaviour is limited and equivocal. Further research is required, utilising precise outcome measurement

Integrating the evidence

There is now robust evidence of associations between self-injury and: repetitive behaviour, health conditions associated with pain, child characteristics (specifically impulsivity, genetic syndromes, ASD) and environmental events to propose that these associations should be accounted for in existing models of the development of self-injury. In Guess and Carr's (1991) model of the development of self-injury, the first stage is characterised by the emergence of rhythmic repetitive behaviours. In Stage Two, these repetitive behaviours function to optimise arousal. During Stage Three, these behaviours become sensitive to environmental (social) reinforcement and are shaped into increasingly severe behaviour. Whilst Guess and Carr's model explains the development of self-injury, it does not account for the elevated prevalence of self-injury in ASD or genetic disorders, and the associations between self-injury and painful health problems, repetitive behaviours and the hypothesised impaired behavioural control.

A revised model of self-injury

On the basis of existing evidence we propose revisions to the Guess and Carr model by identifying a fourth stage and modifying the original three stages. Stage 2 is extended to include behaviours becoming sensitive to all internal states, allowing for these behaviours to have the function of terminating a painful stimulus via pain gating or an attempt by the child to remove the perceived source of the pain. We propose a fourth stage to account for more severe self-injury, in which environmental social control is less influential and self-injurious behaviour is no longer wholly within the individual's control. During this stage, self-restraint behaviours become evident as an attempt to control self-injurious behaviour. In the diagrammatic presentation of this model in Figure 1, a baseline trajectory for the development of self-injury is plotted in accordance with Guess and Carr's (1991) model. Repetitive behaviours occur and then become sensitive to internal states. Over time, these behaviours increase in severity and probability as they are selectively shaped by the environment. The level of external

social control over these behaviours increases from low to high as environmental reinforcement becomes more consistent within a mutual reinforcement paradigm (Oliver et al., 2005).

++++++++++++ Figure 1 here ++++++++++++

A differing trajectory for the development of self-injury for children with characteristics of ASD and specific genetic disorders is proposed. High levels of repetitive behaviour are seen in ASD and some genetic disorders (Estes *et al.,* 2011; Moss, Oliver, Arron, Burbidge, & Berg, 2009; Richler, Bishop, Kleinke & Lord, 2007; Turner, 1999), consequently during Stage One the probability of behaviour and consequently the level of the trajectory of development are elevated. This heightened trajectory remains stable during Stage Two as the behaviours become regulated automatically by internal states. In Stage Three, as social\environmental reinforcement shapes the behaviour, the probability of self-injury in any typical environment to which individuals with relevant phenotypic characteristics are exposed is further heightened, due to phenotype x environment interactions Influential establishing operations and antecedents are hypothesised to occur more frequently for individuals with, for example, ASD impairments and\or genetic syndromes. For example, in a syndrome such as Smith-Magenis, in which adult attention is frequently sought (Wilde et al., 2013) momentary decreases in the level of attention might occasion attention maintained episodes of self-injury (Taylor and Oliver, 2008).

In contrast to ASD and syndrome related motivational variables, it is hypothesised that painful health conditions do not influence the trajectory of self-injury. Instead painful health conditions provide a second pathway for self-injurious behaviour to directly enter the behavioural repertoire in Stage Two. The painful health conditions lead to children engaging in behaviour in an attempt to remove or 'gate' the painful experience. Thus, the starting point of the trajectory for the development of self-injury is higher. The effect of painful health conditions is proposed to be intermittent throughout development, as painful health conditions may occur acutely and then remit. Once established in the behavioural repertoire, these behaviours can be shaped by the environment in Stage Three. Analogous to phenotype x environment interactions, it is hypothesised that the presence of pain also increases the probability of self-injury by

interacting with environmental antecedents to increase social motivation. Therefore, painful health conditions in Stage Three of the model are hypothesised to increase the trajectory of self-injury.

A final person characteristic which the model must account for is the potential influence of impaired behavioural control and apparent absence of social influence. This is hypothesised to affect the development of self-injury at all stages of the model. In Stage One, impaired behavioural control would lead to a heightened prevalence of repetitive behaviours, During Stage Two, behaviour regulates internal states and this prepotent response becomes increasingly difficult for the individual to inhibit. Therefore, the probability *and* trajectory of self-injury are elevated in individuals with impaired behavioural control during Stage Two. Similarly, during Stage Three, prepotent responses to environmental antecedents are difficult to inhibit and thus self-injurious behaviour is initiated more frequently. It is hypothesised that for individuals with impaired behavioural control, it eventually becomes impossible to fully inhibit these prepotent responses and the individual gradually loses control over their self-injury. At this stage, there is transition into Stage Four and the development of self-restraint. For these individuals it is proposed that whilst environmental contingencies may still be active, self-injurious behaviour is no longer wholly controlled by these contingencies. The developmental trajectory for individuals with impaired behavioural control is therefore steepest and of greatest concern.

This revised model demonstrates how children can accrue risk markers which alter the initial probability and developmental trajectory of self-injury. From this, it can be seen that individuals with 'ASD' impairments *and* painful health conditions *and* impaired behavioural control (features that often co-occur in genetic syndromes such as Cornelia de Lange syndrome) may evidence the highest probability of self-injury and the steepest gradient. For these individuals, repetitive behaviours in Stage One are more likely, self-injury can develop in Stage Two via two pathways (repetitive behaviour and health problems), influential environmental antecedents are more likely to be experienced in Stage Three and these individuals are most at risk of progressing into Stage Four during which self-injury is no longer under environmental control.

Clinical implications: Strategic and responsive intervention

The model outlined above has clear implications for interventions. Transition through the stages is associated with increasing severity, prevalence rises with age and self-injury does not resolve without intervention. Consequently, early intervention in childhood is likely to prove a valuable strategic intervention (see Richman, 2008). Additionally, some of the child characteristics associated with self-injury clearly precede the onset of clinically significant self-injury. Genetic syndromes are identified at a very early stage and profound or severe intellectual disability, repetitive behaviour and ASD are likely to be identified in the early years. Impulsivity may be more difficult to establish in the presence of intellectual disability and ASD in young children but appropriate assessment instruments are becoming available. Given the persistence of self-injury, and the observation that most of these child characteristics are typically evident before the age at which clinically significant self-injury emerges, these characteristics might be considered as potential risk markers for future clinically significant self-injury. Clearly, longitudinal data are needed to establish if these are risk markers and their interrelationship but the presence of these potential markers should alert clinicians to this possibility. The identification of high risk children, who accumulate a number of these putative risk markers, is also important as interventions when children progress to Stage 4 are likely to be more difficult. These observations suggest that risk related early intervention is both possible and likely to prove beneficial. Table 1 summarises the potential risk markers for self-injury together with an indication of their contribution to risk.

++++++++++ Table 1 here +++++++++++++++

A second implication is that different interventions are likely to be effective at different stages. Early on, identification and relief of pain or reduction of body contact stereotypies in high risk children is indicated. As the behaviour becomes socially reinforced, functional communication training in combination with contemporary methods of behaviour management are likely to be helpful. Given the phenotype x environment interactions described, interventions would need to be sensitive to child characteristics that might influence motivation. These may be ASD or syndrome sensitive. At Stage 4 when self-restraint and the preference for imposed restraint is evident, the most common methods of experimental functional analysis are frequently

impossible and fading of restraints is more likely to be helpful. This kind of intervention has implications for policies that do not allow use of restraints. It is also clear that movement between the stages might be prevented by anticipating which kind of intervention would prevent further escalation. The pre-emptory use of functional communication training to reduce the possibility of moving from Stage 2 to 3 is a possibility. The final implication is that combinations of interventions might also be warranted. An intervention that addresses compromised behavioural inhibition alongside contingency management and functional displacement might be more effective than either intervention alone in children who are impulsive or who show high rates of repetitive behaviour (see Zarcone et al, 2004 for an interesting demonstration of this.). Table 2 gives some examples of areas of importance for assessment and intervention at each stage of the model.

++++++++++++++ Insert Table 2 here++++++++++++

Research implications

There are also clear implications for research from the proposed model. Longitudinal studies are needed to confirm whether the trajectories of development of self-injury do differ with child characteristics and whether the stages are each necessary or, for example, whether it is possible to move from Stage 2 to 4 without social reinforcement. It is also interesting to consider whether phenotype x environment interactions predicted on the basis of the presence of, for example, syndrome or ASD diagnosis might reduce the need for standard experimental functional analysis or modify the order of stimuli assessed. Finally, the utility of randomised controlled trials of self-injury that do not take into account cause is questionable. If intervention trials have self-injury alone as the inclusion criterion and do not attend to child characteristics or behavioural correlates of self-injury and do not assess cause then the group result will be substantially affected.

Research priorities include the identification and perception of pain in children who cannot self-report, increasing the efficiency of assessment strategies whilst maintaining robust reliability and validity and integrating assessment strategies so that case study and group design intervention reports comment on pain, behavioural correlates such as impulsivity, self-restraint, ASD, genetic syndromes regardless of the nature of the

intervention. Similarly, the properties of observed self-injury (temporal patterns, from simultaneously or closely occurring behaviours) that might be associated with different stages of the proposed model warrant description to aid assessment. Finally, the association between self-injury and genetic syndromes should be further explored, particularly the stable topography of self-injury seen in some syndromes, despite environmental influences. The potential group contrast designs are strong methodologically and allow greater control over confounding variables.

Barriers to implementation

The most important clinical issues are the use of medication without supporting evidence or systematic evaluation, the use of non-evidenced based psychological interventions such as psychodynamic therapies and the lack of the provision of applied behaviour analytic interventions when these are clearly indicated (see Ruddick et al., in review). It is not clear why there is such a widespread failure in services to deliver demonstrably effective interventions based on applied behaviour analysis that are supported by such a strong empirical literature. One influence is clearly the widespread and persistent failure of clinical psychology training to respond to the level of clinical need and a review of the provision of this service delivery is warranted. Similarly, training for other multi-disciplinary professionals in psychiatry, education and social care must improve to include some recognition of the efficacy of behavioural interventions, and the necessity of applied behaviour assessments for self-injury.

An additional barrier to implementation is the current reactive nature of clinical services. Support and intervention are rarely offered until the behaviour has become entrenched and costly for both the individual and families/services. At this point, it is difficult and sometimes dangerous to implement behavioural interventions which may have been effective earlier when behaviour was less severe and the learning history shorter. The research literature reviewed above highlights those characteristics which make an individual more likely to develop self-injury, and the revised model presented identifiable characteristics which are associated with more severe self-injury. The ability to identify ‘at risk’ individuals and groups could lead to an alternative service structure, informed by a model of early intervention wherein a more pro-active and putatively more effective stance on intervention could be taken. However, whilst services focus on only those with the most severe and entrenched behaviour, it is

perhaps inevitable that interventions for self-injury will continue to be reactive and limited in efficacy.

Key practitioner message

- Self-injury is common in individuals with intellectual disability. The behaviour is persistent without effective intervention and has a significant impact upon quality of life.
- Operant models of self-injurious behaviour can account for the development and maintenance of self-injury in many cases. Functional interventions derived from applied behaviour analysis are indicated in most cases.
- Individual characteristics, specifically the presence of autism, certain genetic syndromes, painful health conditions, repetitive behaviour and impulsive behaviours are associated with an increased risk of self-injurious behaviour.
- These risk markers add to an understanding of self-injury that builds upon the operant model to allow for more sophisticated phenotype x environment interactions.
- Limited practitioner training in effective assessment and intervention for self-injury, and the reactive focus of services prevents clinical improvements for self-injurious behaviour.

Areas for future research

- Longitudinal studies evaluating the development of self-injury relative to individual characteristics, specifically painful health conditions, repetitive behaviours and behaviours indicative of behaviour dysregulation.
- Randomised control trials for interventions for self-injury that take into account differing causes for the behaviour.
- Further investigation of phenotype x environment interactions and delineation of the associations between specific genetic syndromes and self-injury.
- Improving tools to identify pain in individuals who cannot self-report.

Clinical commentary

Self-injury is a common and intransigent behaviour for many individuals with intellectual disability. Operant learning models and their derived interventions continue to be the most effective and evidence based approaches; however, they are often neglected in clinical practice. This paper presents research delineating phenotype x environment interactions and suggests a developed model of self-injury to account for

these interactions. Specifically, the influence of pain upon self-injury is discussed. Additionally, the influence of impaired behavioural control on the development of self-injury is hypothesised.

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Table 1. Summary of putative risk markers for self-injury from empirical research studies with corresponding descriptions of the risk marker and odds ratio.

Putative Risk Marker	Description of putative risk marker	Odds Ratio (CI)
Degree of Intellectual Disability	Meta-analysis using varied criteria	4.06 (2.56-6.43) a
	More severe deficit in adaptive behaviour on a measure of self-help skills	3.15 (CI not reported)[d]
	The presence of lower levels of ability on a standardised measure of self-help skills (child sample)	3.84* (1.60 – 9.19)[f]
	Level of ID (mild, moderate, severe/profound)	2.11 (1.64 – 2.72) [g]
	Severe/profound ID vs mild/moderate	7.19 (3.27–15.82)[i]
Autism	Meta-analysis using varied criteria	5.6 (1.39-22.56) [a]
	Meeting criteria for autism on a standardised measure	2.67[+] (1.45-4.91)[c]
	Diagnosis of autism	1.70 (1.03–2.80)[g]
Genetic Syndromes	Cri du Chat syndrome	9.04 (2.93-27.88) [b]
	Cornelia de Lange syndrome	6.47 (2.48-16.86) [b]
	Fragile X syndrome	2.88 (1.22-6.82) [b]
	Prader Willi syndrome	2.91 (1.23-6.91) [b]
	Lowe syndrome	4.92 (1.71-14.17) [b]
	Smith Magenis syndrome	35.53 (6.32-199.92) [b]
		0.24 (0.055–0.997)[i]
	Down syndrome	0.36 (0.20 – 0.64)[g]
Repetitive/Stereotyped Behaviour	The presence of high frequency repetitive or ritualistic behaviour	6.43 (CI not reported)[d]
	The presence of high levels of repetitive and stereotyped behaviour (adult sample)	2.57 (1.04 – 6.39)[f]
	The presence of stereotyped behaviour on a standardised measure	0.23*[h]
	The presence of high levels of repetitive and stereotyped behaviour	2.66 (1.84, 6.02)[e^]
Health Problems	The presence of one or more health problems (child sample)	3.54 (1.49 – 8.40)[f]
	Visual impairment	1.94 (1.01–3.72)[i]
Overactive/Impulsive Behaviour	The presence of high levels of overactive and impulsive behaviour (child sample)	5.71 (2.22 – 14.72)[f]
	The presence of high levels of overactive and impulsive behaviour (adult sample)	3.92 (1.72 – 8.95)[f]
	The presence of impulsive behaviour on a standardised measure	0.46*[h]
	Meeting criteria for ADHD on a standardised measure	10.95 (3.50–34.19)[i]
Sensory sensitivity	Tactile hypersensitivity	2.23 (1.23–4.04)[g]

McClintock, Hall, & Oliver (2003)[a]; Arron, K., Oliver, C., Moss, J., Berg, K., & Burbidge, C. (2011)[b]; Richards, Oliver, Nelson, & Moss (2012) [c]; Oliver, C., Petty, J., Ruddick, L., & Bacarese-Hamilton, M. (2012) [d]; Davies & Oliver (2014) [e]; Richards, Davies & Oliver (In Review)[f]; Lundqvist (2012)[g]; Richman, Barnard-Brak, Bosch, Thompson, Grubb & Abby (2012) [h]; Cooper *et al.*, 2009[i]

[+] Compared to those with Down syndrome
[*] Standardised path value
^ This relative risk reflects the onset of self-injury associated with the putative risk marker, rather than the presence of self-injury

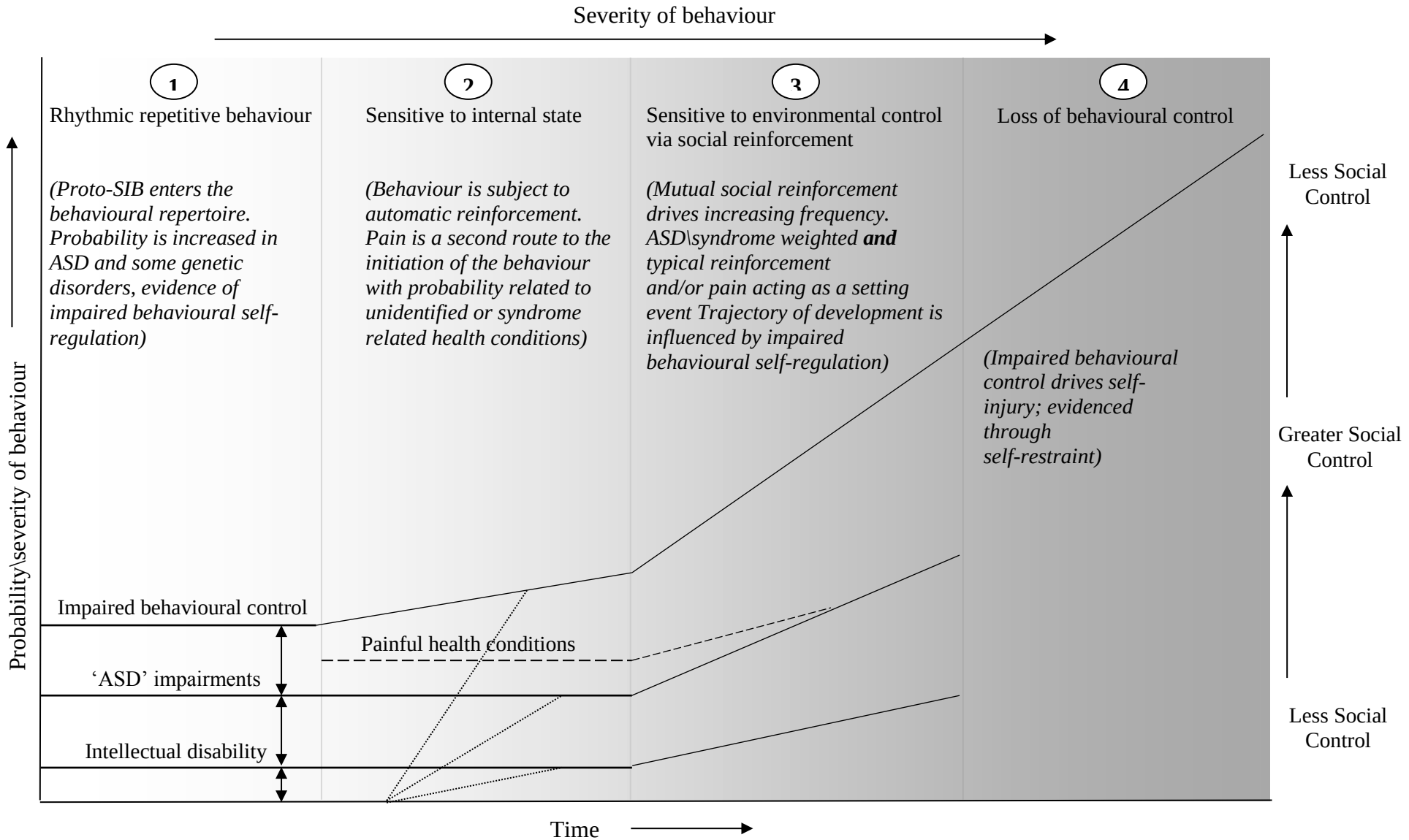

Figure 1 Hypothesised model of the development and maintenance of self-injury in ASD over time (see text).

Table 2. Areas of focus for assessment and intervention at each stage of the proposed model.

Stage	Assessment	Intervention
One: Rhythmic, repetitive behaviour	• Assessment of person characteristics to identify those children with multiple 'risk markers' in order to target proactive early intervention.	• Broad communication interventions to improve a functional communication. • Documentation of a child's 'typical' behaviour when healthy; accumulation of observations of a 'pain signature'.
Two: Sensitive to internal states	• Regular medical assessment of physical health problems. Any significant changes in behaviour should prompt a reassessment to rule out untreated pain and discomfort. • Assessment of body contact stereotypies; include functional assessment.	• Appropriate medical interventions to alleviate pain and painful health conditions. • Where body contact stereotypies are functioning to reduce or increase arousal, alterations to the environment.
Three: Sensitive to environmental control via social reinforcement	• Functional assessment of emerging self-injurious behaviours; experimental functional analysis where appropriate to identify any functions to the behaviour. • Identify idiosyncratic motivational operations relevant to phenotype x environment interactions. • Identify precursor behaviours. • Assess physical health. • Vigilance for emerging self-restraint behaviour.	• Specifically designed communication interventions as a result of functional analysis assessments e.g., Functional Communication Training. • Environmental manipulations to change antecedents or maintaining consequences. • Appropriate medical interventions to alleviate pain and painful health conditions.
Four: Loss of behavioural control	• Assess preference for restraint through brief removal and then replacement of any imposed restraint, assessing the child's affect and attempts to self-restrain or gain access to the imposed restraints.	• Restraint fading.